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Enteropathy-Associated T-Cell Lymphoma: A Case Report With Radiographic and Computed Tomography Appearance

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We report a case of enteropathy-associated T-cell lymphoma (EATL) of the jejunum in a 56-year-old man. The patient suffered for several years from nonspecific abdominal complaints, with no clinical evidence of malabsorption. The patient underwent extensive imaging procedures including barium meal and computed tomography. Computed tomography of the abdomen showed small mesenteric lymph nodes and an area of intestinal wall thickening. Barium meal demonstrated a short jejunal stricture. Histology revealed lymphoma of the jejunum, with microscopic changes distant from the lesion consistent with celiac disease. The spectrum of EATL ranges from patients with frank celiac disease, to patients with only immunohistochemical evidence of celiac disease, who develop small bowel lymphoma. *J. Surg. Oncol. 1997;65:50–54.*

KEY WORDS: lymphoma—small bowel; enteropathy-associated; malabsorption; celiac disease

INTRODUCTION

Enteropathy-associated T-cell lymphoma (EATL) is an unusual malignancy presenting in patients with a malabsorptive state that may be clinically evident or clinically silent with only laboratory evidence of its existence.

CASE REPORT

The patient was a 56-year-old man whose initial symptoms dated back some 10 years with heartburn, upper abdominal pain, vomiting, and weight loss. Radiographic examination of the upper gastrointestinal tract, including small bowel series, and abdominal sonography were reportedly normal. For the next 3 years, the patient received symptomatic treatment and remained relatively well, until 9 months prior to admission, when abdominal pain recurred, accompanied by nausea. There was no diarrhea or evidence of malabsorption. At that time gastroscopy revealed hiatal hernia and changes consistent with duodenitis. He again received symptomatic treatment and was referred for abdominal computed tomography (CT), which showed small mesenteric lymph nodes (Fig. 1). Follow-up CT 6 months later was interpreted as normal. Laboratory examinations were noncontributory.

The current admission was prompted by abdominal pain and vomiting. Gastroscopy showed only hiatal hernia. Endoscopic retrograde cholangiopancreatography (ERCP) was normal. Small bowel series showed a focal narrowing in the jejunum (Fig. 2) with mild dilation of proximal loops. Reviewing the previous CT examination, an area of small bowel narrowing was noted retrospectively (Fig. 3).

The patient underwent laparotomy with resection of his jejunal tumor and nearby omentum, and end-to-end anastomosis. There were enlarged regional lymph nodes. Despite chemotherapy, the patient died of extensive disseminated lymphoma 6 months later.

PATHOLOGY

The resected fragment of the small bowel measured 28 cm and included nearby omentum. At a single location there was found a constrictive lesion formed by white

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Fig. 1. Axial CT image through the mid-abdomen showing small mesenteric lymph nodes.

tumor tissue 5 cm in diameter, which penetrated the bowel (Fig. 4). The overlying mucosa appeared to be edematous and partially ulcerated. The proximal portion of the small bowel was dilated. Enlarged lymph nodes were found in the omentum.

On histological examination, the tumor consisted of large anaplastic cells with pleomorphic nuclei and prominent nucleoli (Fig. 5). Those cells were similar to Reed-Sternberg cells and were admixed with mononuclear cells. Lymphatic permeation, as well as diffuse infiltration of the bowel and surrounding fatty tissue were prominent features. The enlarged lymph nodes showed infiltration by the tumor cells.

An additional histologic feature was the presence of celiac-like changes in the small bowel mucosa lying a distance from the tumor (Fig. 6).

The immunohistochemical results showed positive staining of the malignant cells with Vimentin, LCA, UCHL 1 (T-cell marker), BER-H2 and also EMA. Based on the findings, a diagnosis of anaplastic large T-cell lymphoma (K-1 positive) with mucosal associated celiac-like changes was made.

DISCUSSION

The gastrointestinal tract is the most common site for primary extranodal lymphomas. The small intestine accounts for 37% of these tumors, which characteristically involve the ileum more frequently than the jejunum. With the advent of immunohistochemical markers, lymphomas have been classified as either T-cell or B-cell variety. In one series of 119 cases of primary small intestinal lymphoma collected over a period of four decades [1], 66% were B-cell and 34% T-cell tumors. Eighty-three percent of the T-cell tumors were high-grade malignancies, and 49% were enteropathy associated. The prognosis of these tumors varies greatly, with five-year survival ranging from 75% for low-grade B-cell tumors to only 25% for T-cell tumors. In another series, of 27 T-cell lymphomas of the intestine, 92% were confined to intestine and abdominal lymph nodes, and 33% were enteropathy associated [2].

The first published accounts of the association of gastrointestinal lymphoma and malabsorption attributed the malabsorption directly to the lymphoma [3]; however,

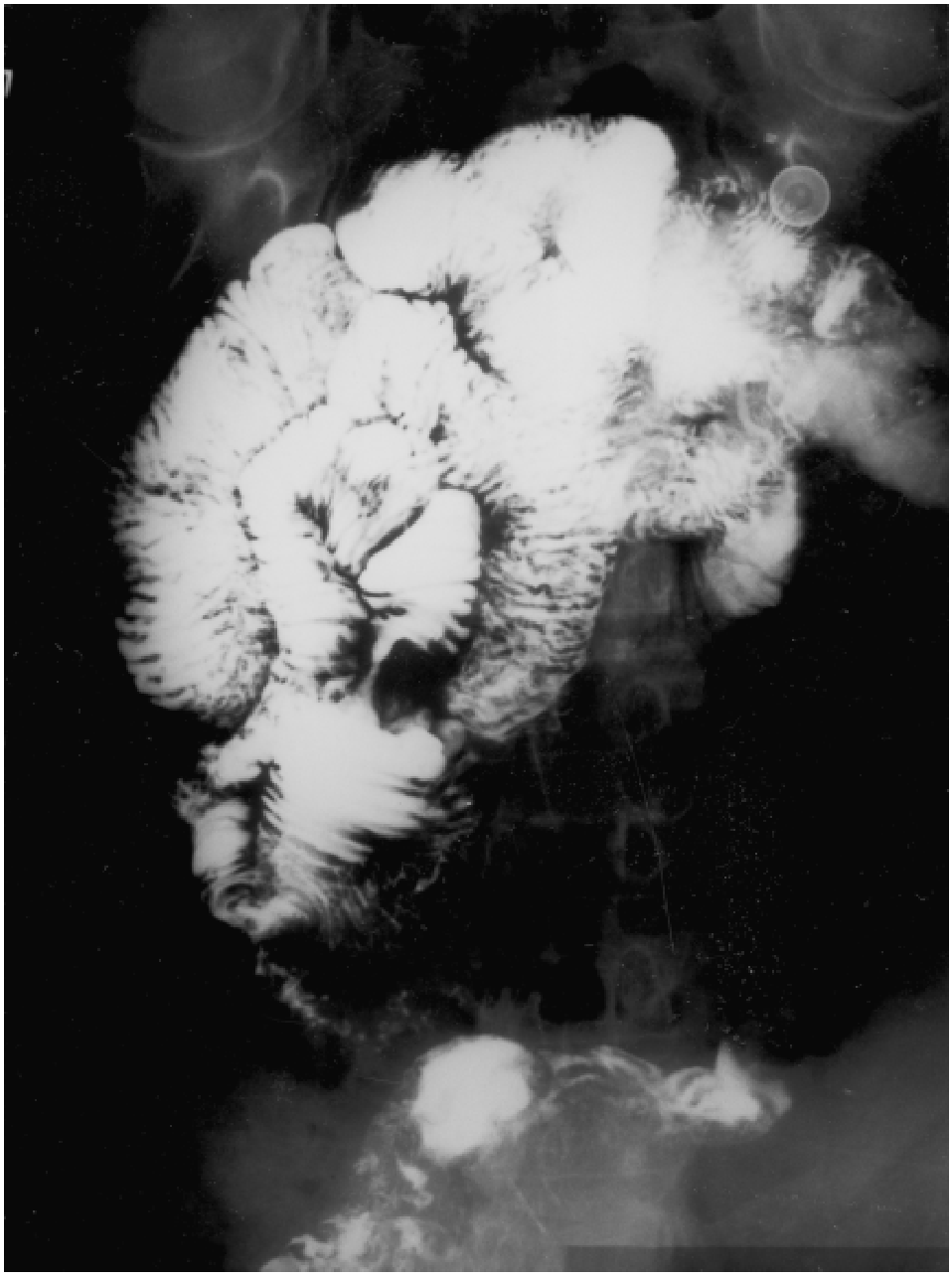

Fig. 2. Constricting lesion on small bowel barium meal examination.

malabsorption may precede the diagnosis of lymphoma by several years [4]. The association of malabsorption and lymphoma occurs across a clinical spectrum: in patients with frank celiac disease; in patients with intestinal mucosal pathology without evidence of malabsorption; or in patients with latent celiac disease, in whom there is abnormal intestinal mucosal immunity without histologic evidence of villous flattening [5,6].

Immunohistochemical and molecular studies have shown these lymphomas to be of T-cell origin [7] and they are currently termed enteropathy-associated T-cell lymphoma. The tumor apparently originates from intraepithelial lymphocytes in the small intestine, and may stay localized for a prolonged period without evidence of extraintestinal spread.

The median age at diagnosis of EATL is 60 years, and there is a slight male preponderance. This tumor usually affects the jejunum, at times in combination with other sites in the gastrointestinal tract, and on gross examination takes the form of circumferential ulcers without the formation of large tumor masses. Mesenteric lymph node enlargement is characteristic. The patient may have suf-

Fig. 3. Axial CT image through the lower abdomen showing a narrowed bowel loop with thickened walls.

Fig. 4. The resected fragment of small intestine showing the tumoral constriction.

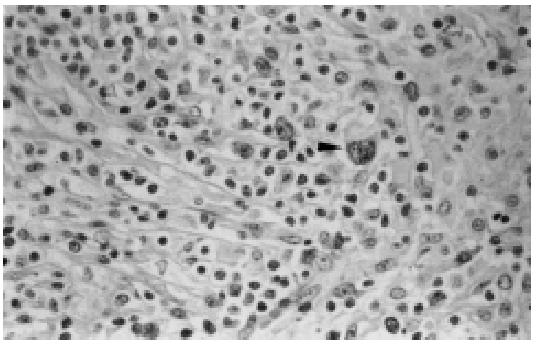

Fig. 5. Prominent anaplastic Reed-Sternberg-like cells (arrowheads) admixed with mononuclear cells. Hematoxylin and eosin, ×40.

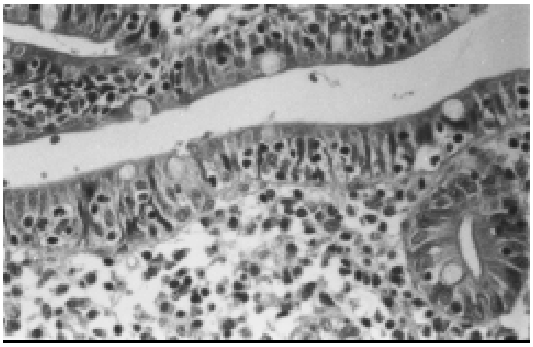

Fig. 6. Celiac-like changes of the small intestinal mucosa with lymphocytic infiltration and disappearance of villi.

fered from abdominal pain, weight loss, and nonspecific symptoms for a period of years; however, presentation of this disease as an acute illness with perforation, obstruction, or hemorrhage is not uncommon. A small proportion of patients have a history of celiac disease dating back to childhood. In many cases of EATL, there is no history of celiac disease or biopsy evidence of gluten sensitivity, and the resected jejunal mucosa may appear normal.

Malignant small bowel lymphoma has a wide variety of radiologic manifestations. Ulceration, narrowing, polypoid mass, mechanical obstruction, intussusception, fistulas, aneurysmal bowel dilation, thick mucosal folds, separation of adjacent loops, mesenteric adenopathy, and mesenteric thickening are all among the frequent imaging findings on barium meal examination and CT [8]. Most T-cell tumors of the small intestine present as ulcerated plaques or strictures in the proximal small bowel, while B-cell tumors tend to be annular or polypoid masses in the distal or terminal ileum [1].

In this case report, our patient presented with nonspecific abdominal complaints for a number of years prior to developing frank malignancy. Malabsorption was not an evident clinical problem. The finding of a tumoral stricture in the proximal small bowel was consistent with the subsequent diagnosis of EATL.

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